CASE REPORT

Isolated laryngeal amyloidosis in a patient with a hoarse voice

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SUMMARY

We present a case of a 59-year-old woman with a hoarse voice of 3 weeks duration. A polypoid lesion involving the anterior aspect of the left vocal cord was detected by flexible fiberoptic endoscopic examination. Flexible bronchoscopic examination was unremarkable. Microlaryngoscopy and excisional biopsy of the laryngeal lesion were carried out. Histology of the biopsy showed stroma exhibiting birefringence under polarised light and a diagnosis of laryngotracheal amyloidosis was made. Systemic amyloidosis was ruled out by further investigations including positron emission tomography scan. Her hoarse voice improved following the excisional biopsy and regular follow-up was arranged for early diagnosis of recurrence.

BACKGROUND

Localised amyloidosis in the head and neck is a rare benign disease that accounts for 0.2–1.5% of benign laryngeal tumours. Amyloidosis is diagnosed by Congo red staining demonstrating the classic appearance of bright green birefringence under polarising light microscopy. The most effective treatment for local laryngeal amyloidosis is microlaryngoscopy with excision. The prognosis for laryngeal amyloidosis is usually good. Regular follow-up is important for early detection of recurrence owing to the significant likelihood of recurrent or residual disease. This case presents an unusual cause of a hoarse voice and highlights diagnostic approach and management issues relating to localised amyloidosis.

CASE PRESENTATION

A 59-year-old woman presented with a worsening hoarse voice of 3 weeks duration. Her medical history was unremarkable and she was not on any regular medications. She had been smoking approximately 5–10 cigarettes for the past 30 years. There was no history of trauma, recent intubation, upper respiratory tract symptoms, dyspnoea or stridor. She denied any weight loss; night sweats or any head and neck mass. She also denied any symptoms possibly pointing to hypothyroidism. Flexible nasal endoscopic examination of the larynx showed a polypoid lesion involving the anterior aspect of the left vocal cord. The rest of clinical examination was unremarkable.

INVESTIGATIONS

Microlaryngoscopy was carried out which confirmed the flexible nasal endoscopic examination findings. Flexible bronchoscopy did not show

Figure 1 Micrograph showing laryngeal amyloid deposition (H&E original magnification $\times 20$).

tracheal involvement. Histology of the biopsy taken during flexible bronchoscopy revealed a stroma expanded by an amorphous eosinophilic material (figure 1) that was positive with Congo red staining (figure 2) and exhibited birefringence under polarised light.

A diagnosis of laryngeal amyloidosis was made. Further investigations were carried to rule out systemic amyloidosis. The patient's complete blood count, erythrocyte sedimentation rate, basic metabolic and biochemical panel and liver function tests were within normal limits. Serum calcium as well as serum and urine electrophoresis were also normal. Positron emission tomography scan showed mildly fluorodeoxy-D-glucose-avid left vocal cord but did not show any other abnormalities.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of a hoarse voice is broad including benign pathological entities of acute viral

Figure 2 Micrograph showing laryngeal amyloid deposition (Congo red original magnification $\times 20$).

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laryngitis and postnasal drip. Systemic processes including hypothyroidism can also cause a hoarse voice. Possible compressive effect on the recurrent laryngeal nerve also needs to be considered. Entities such as bronchogenic carcinoma and malignant oesophageal tumours need to be ruled out. Neurological causes of hoarseness include stroke, Parkinson's disease, myasthenia gravis, multiple sclerosis and amyotrophic lateral sclerosis, can be responsible for hoarseness of voice. Once a nodule was identified in the larynx/trachea, possible diagnoses narrow. They include primary laryngeal carcinoma, papilloma, polyps and cysts. If the vocal cord is involved diffusely, Reinke's oedema needs to be considered.

TREATMENT

Her hoarse voice improved significantly following the excisional biopsy.

OUTCOME AND FOLLOW-UP

Regular 3-month follow-up appointments have been arranged for early diagnosis of recurrence for at least 5 years.

DISCUSSION

Amyloidosis is a disorder caused by the extracellular deposition of normally soluble proteins in an abnormal fibrillar form. An excess of 20 proteins are known to form insoluble fibrils in humans.¹ They present amorphous eosinophilic appearance under light microscopy following H&E staining and bright green fluorescence observed under polarised light after Congo red staining.¹ Amyloid is classified histochemically.¹ In formerly called primary amyloidosis and myeloma-associated amyloidosis, the fibril protein is an immunoglobulin light chain abbreviated as L. These patients, designated amyloid light-chain (AL), are termed to have light chain amyloidosis. In amyloid A (AA-secondary amyloidosis), harmful deposits of amyloid are produced because of various inflammatory diseases.¹

Localised amyloidosis in the head and neck is a rare benign disease.² The larynx is the most common site of involvement and accounts for 0.2–1.5% of benign laryngeal tumours.³ It usually affects individuals in the 40–60 years age range, with a male-to-female predominance of 2:1.⁴ The disease runs a slowly progressive course and is characterised by hoarseness, cough, dyspnoea and less commonly stridor. Clinically, the lesion most commonly appears as a firm and non-ulcerated yellowish submucosal nodule or polypoid lesion.⁴ Within the larynx, the ventricles and vestibular folds are most commonly involved.⁴

Once systemic amyloidosis is ruled out biochemically and radiologically, treatment of choice is endoscopic excision.² Cold resection of amyloid lesions may be complicated by bleeding

owing to the tendency of amyloid to infiltrate local blood vessels.⁵ Carbon dioxide laser excision may be advantageous in very extensive lesions. However, its use may also be complicated by extensive postoperative scarring.⁵ Local or systemic steroids are found to be ineffective in the treatment of localised laryngeal amyloidosis.⁴

Although the prognosis for laryngeal amyloidosis is usually good, the likelihood of recurrent or residual disease is significant and death from pulmonary failure has been reported.⁶

Regular follow-up is important for early detection of recurrence. Relapse may occur after a long time period; long-term follow-up is essential for at least 5–7 years.⁴ A high degree of suspicion is necessary since clinical presentation of the disease may imitate that of a laryngeal neoplasm.

We present a case that underlines the role of birefringence from Congo red stain to confirm the diagnosis and management strategy.

Learning points

- ▶ Localised amyloidosis is a rare benign disease affecting the larynx.
- ▶ Diagnosis is made histochemically with characteristic positivity with Congo red staining and birefringence under polarised light and once systemic amyloidosis is ruled out.
- ▶ Treatment for local laryngeal amyloidosis is endoscopic excision.
- ▶ Follow-up is essential owing to significant likelihood of recurrence.

Competing interests None.

Patient consent Obtained.

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